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A case report

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Melena as presentation of primary small intestine inflammatory myofibroblastic tumor in an adult woman. A case report

INTRODUCTION: *Inflammatory myofibroblastic tumor (IMT), also known as inflammatory pseudotumor, plasma cell granuloma or inflammatory myofibroblastoma, is characterized histopathologically by myofibroblastic spindle cells with inflammatory cell infiltrates (plasma cell, lymphocytes and eosinophils). Inflammatory myofibroblastic tumor is typically seen in children or young adults and is most commonly localized in the lung, but it can occur anywhere in the body.*

CASE REPORT: *We present a case in a young woman with severe acute G.I. bleeding, an uncommon presentation of IMT in adults. The patient was admitted to the emergency department for melena.*

MRI showed a distal jejunum hypervascular mass. Other exams were negative. Surgical excision was recommended, so the patient underwent surgery with complete removal of the tumoral mass. No adjuvant therapy was employed and the patient is asymptomatic after 6 months of follow-up.

DISCUSSION: *IMT is an rare lesion that mimics malignancy and is accompanied by various clinical manifestations. The treatment of choice is believed to be complete surgical excision and long term follow up.*

KEY WORDS: Anemia, Inflammatory myofibroblastic tumor (IMT), Small intestine, Surgery

Introduction

Inflammatory myofibroblastic tumor (IMT), inflammatory pseudotumor and plasma cell granuloma are terms that have been used interchangeably for a variety of lesion ranging from some that are inflammatory for nature, to those that are clearly neoplastic. Although neo-

plastic lesion generally pursue a benign clinical course, intraabdominal and retroperitoneal lesions of IMT. have typically shown higher local recurrence rates and even distant metastases. Lesion have been reported in a variety of intraabdominal organs, but small bowel tumors are particularly rare.

Case Report

A 32 year-old female was admitted to the first aid department with melena and tachycardia, headache and asthenia.

Her past medical history was characterized by laparoscopic removal of ovarian cyst 5 year ago.

Clinical examination did not reveal any palpable abdominal mass and there were no sign of tenderness. Routine

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blood tests revealed hemoglobin level of 5.8 g/dl so she received a transfusion of two units of blood. Gastroscopy was negative, sigmoidoscopy didn't show any suspicious lesion of the colon, chest x – ray was negative. Because of the persistence of melena the patient was admitted to our emergency department, routine blood test revealed hemoglobin level of 7.6 g/dl, the MRI showed a parietal thickening involving the medium distal jejunum, with a 20.0 x 20.0 mm hypervascular mass. A diagnosis of neuroendocrine tumor or a GastroIntestinal Stromal Tumor was suspected; serum Chromogranin A and neuron-specific enolase (NSE) were negative. Surgical excision was recommended. The patient underwent exploratory laparotomy; tumor's location was confirmed 30 cm distally to Treitz ligament. The lesion was a small 2 cm diameter hypervascular mass; no lesions or lymph nodes enlargement were evident. The final histopathology reported an intussusception of small bowel tract with parietal thickening due to a myxoid like lesion with a central ulcer of 1.6 cm diameter. Microscopic examination of surgical sample revealed a proliferation of loosely arranged plump and spindled myofibroblasts in an oedematous myxoid background with a mixture of inflammatory cells. The immunohistochemistry technique showed positivity for smooth muscle actin and negativity for DOG1, S100, CD117 and CD34. Immunological and morphological findings were found to be consistent with the diagnosis of IMT. The post-operative course was uneventful and the patient appears to be doing well over 6 months of follow-up with normal hemoglobin level.

Discussion

IMT is a rare, but distinctive spindle cell tumor that contains a variable number of inflammatory cells, including plasma cells. This is the reason for previous designation of mass as plasma cell granuloma, but plasma cell granuloma as well as other alternative terms are discouraged to be used (including inflammatory pseudotumor, inflammatory myofibroblastoma and inflammatory myofibrohistiocytic proliferation)¹⁻⁵. Characteristic histopathological findings in typical IMT are a fasciitis-like, compact spindle cell proliferation with areas of myxoid change and hypocellularity showing a collagenous background⁶. Various numbers of mixed inflammatory cells including polyclonal plasma cells, lymphocytes, eosinophils and rarely foamy macrophages are invariably seen^{7,8}. Typically the spindle cells of the IMT express vimentin, smooth muscle actin and other markers which correspond to the myofibroblastic nature of these cells⁸⁻¹⁰. It is important to take in account that while definitive histology has characteristic features, bioptic sampling of the lesion that may lead to interpretation problem since IMT may simulate granulation tissue or a reactive process. Histological features usually cannot predict the

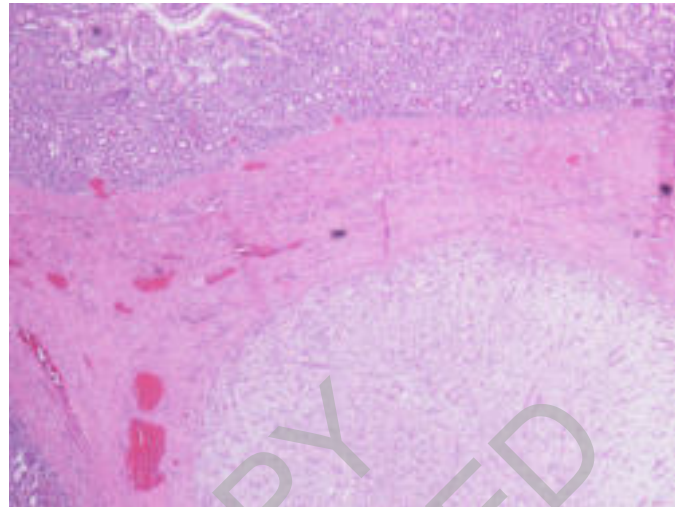


Fig. 1: Lesion within the jejunal wall (E/O).

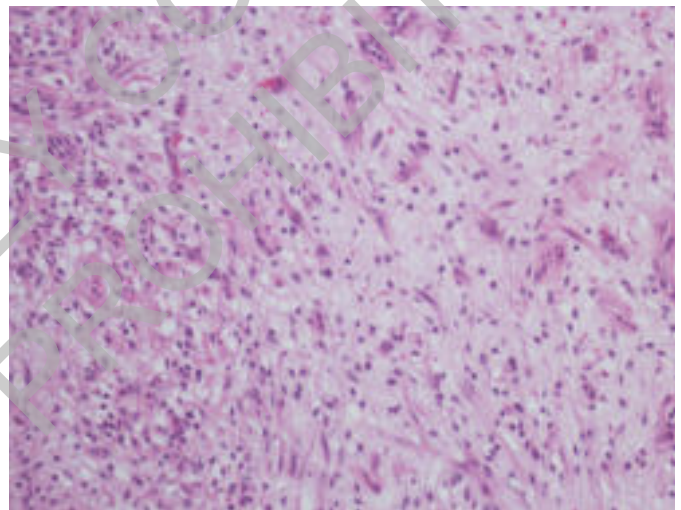


Fig. 2: Details of the lesion (E/O).

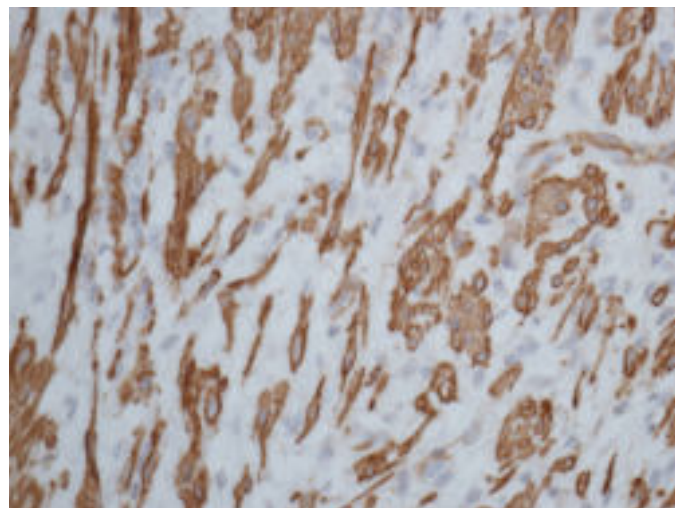


Fig. 3: Immunohistochemistry staining of the lesion for smooth muscle actin (SMA).

biological behavior of the tumor. The presence of aneuploidy may, however, indicate the possibility of a local aggressive behavior and recurrence¹¹. In pulmonary lesions atypia of the spindle cells is believed to indicate aggressiveness.⁸ IMT is more frequent in children and young adults but can develop in older ages with no predilection for any sex^{2,7,10,12}. The lung is the most common site of involvement but IMT lesion have also been reported in other organs and sites including stomach, mesentery, omentum, retroperitoneum as well as kidney, renal pelvis, liver, spleen, esophagus and lymph nodes^{4,5,7,13,14}. IMT has a well known tendency for local invasive behavior and recurrence^{2,3,15}. Only a small risk of distant metastasis has been reported by some authors⁵⁻⁸. The treatment of choice is believed to be complete surgical excision and long term follow up with physical examination, imaging tests and serial monitoring of the erythrocyte sedimentation rate¹⁶. When the mass is assessed as unresectable by a CT scan or laparoscopic exploration, surgical procedures should be avoided and conservative therapy with antibiotics, steroids, NSAIDs, or observation along with intense follow-up should be taken into consideration¹⁷. Intussusceptions in adults always denote intraparietal pathology as was the case in our patient. The patient underwent surgery with complete removal of the obstructing tumoral mass. No adjuvant therapy was employed and the patient is asymptomatic after 6 months of follow-up without any evidence of acute or chronic GI bleeding and normal hemoglobin level.

Conclusions

In conclusion, IMT is a rare lesion that mimics malignancy and is accompanied by various clinical manifestations; an IMT must be considered as the possible cause of subocclusion and melena.

Riassunto

È il caso di una paziente di 32 anni affetta da Miofibroblastoma non noto, trasferita da altra struttura sanitaria presso il nostro reparto perché affetta da melena da fonte non determinata.

All'anamnesi risultava un precedente ricovero in altro Pronto Soccorso per un quadro clinico caratterizzato da dispnea, cefalea, tachicardia e astenia profonda, oltre a profonda anemia (Hg 5.8 g/dl). La gastroscopia non aveva evidenziato alcun motivo di sanguinamento e l'ecografia addomino-pelvica era risultata negativa. Dopo ripetute emotrasfusioni la tentata rettosigmoidoscopia era stata interrotta per intolleranza della paziente.

Una volta stabilizzata si era proceduto al trasferimento presso la nostra struttura, e qui alla Risonanza Magnetica dell'addome veniva evidenziato, un ispessimento parietale

le esteso per 8 cm a del digiuno medio distale, in corrispondenza del fianco destro, antero – lateralmente al duodeno, ed adiacente ad esso una formazione ipervascolare solida di 20 mm. In prima ipotesi poteva trattarsi di un tratto invaginato del piccolo intestino con all'apice una lesione parietale solida, verosimile causa del sanguinamento. Persistendo il quadro anemico e avendo localizzato una lesione sospetta, la paziente è stata sottoposta ad intervento chirurgico, che portava alla conferma dell'esistenza di una lesione nodulare parietale del piccolo intestino, per cui si procedeva ad una sua resezione segmentaria comprendente la suddetta massa, sottoponendo il pezzo operatorio ad esame istologico definitivo.

L'anatomo-patologo formulava la diagnosi isto-immunologica di pseudotumore infiammatorio (c.d. Tumore miofibroblastico infiammatorio) responsabile di invaginazione digiuno-digiunale. I margini di resezione risultavano indenni.

Dimessa dopo un decorso postoperatorio senza complicanze, al follow-up di 6 mesi non si è ripresentato alcun sintomo di nuovo sanguinamento.

Il sanguinamento della neoformazione responsabile della melena da fonte ignota era dovuto alla ipervascolarizzazione della tumefazione rivelata con la RMN, ed alla soluzione di continuo della mucosa di rivestimento conseguente al trauma della intussuscezione e non semplicemente alla sua struttura istologica.

Si conclude per la remota possibilità di un Tumore Miofibroblastico Infiammatorio può essere responsabile di intussuscezione, come ogni massa parietale dell'intestino, e di melena.

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Commento e Commentary

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La rarità dello pseudotumore infiammatorio miofibroblastico ne fa una entità nota più agli anatomo-patologi in fase postoperatoria che non ai chirurghi nel preoperatorio, per la indeterminatezza dei sintomi classicamente legati a questa entità nosologica: febbre, perdita di peso, aumento della VES, anemia e leucocitosi, aumento delle gamma-globuline e della conta piastrinica.

L'adozione delle moderne tecniche strumentali di imaging, che consentono anche lo studio della vascolarizzazione di strutture anomale, sono la base di diagnosi un tempo raramente realizzabili ad di fuori di riscontri operatori di difficile indicazione. La precisazione di natura di pseudotumore miofibroblastico infiammatorio si basa evidentemente non solo sull'aspetto strutturale istologico ma sullo studio istochimico ed immunologico.

Il fatto che la manifestazione principale del caso descritto fosse una melena a sede indeterminata, è ascrivibile appunto all'ipervascolarizzazione della tumefazione rivelata con la RMN, e non semplicemente alla sua struttura istologica, ben diversa dall'angiomiolipoma, ma soprattutto al fatto che si trattava di una soluzione di continuo della mucosa in corrispondenza dello pseudotumore responsabile di una intussuscezione digiunale. Sorprende che dall'insieme della storia clinica non siano segnalate coliche intestinali legate all'intussuscezione, che si deve presumere che fosse periodica.

* * *

The rarity of inflammatory myofibroblastic pseudotumor makes of it an entity more known postoperatively to pathologists than preoperatively to surgeons because of the vagueness of the symptoms classically associated with this nosologic disease: fever, weight loss, elevated ESR, anemia and leukocytosis, elevated gamma globulins and platelet count.

The adoption of modern techniques of imaging, which allow also the study of the vascularization of abnormal structures, are the basis of diagnosis once rarely feasible outside of final pathologic report from operations done with not sure indication. The clarification of the nature of inflammatory myofibroblastic pseudotumor is evidently based not simply on ordinary histology but on histochemical and immunological analysis of the specimen.

The fact that the main event of the presented case was a melena from unknown source depends on the hypervascolarization of the swelling detected by MRI, and not merely on its histological structure, very different from angiomyolipoma, but above all to the fact that there should be a pressure erosion of the mucosa in correspondence of the pseudotumor responsible for a jejunal intussusception. Surprisingly, in the entire medical history is lacking the report of intestinal colic related to periodic intussusception, presumptively periodic.